American Journal of Ophthalmology Case Reports 25 (2022) 101388

Contents lists available at ScienceDirect

American Journal of Ophthalmology Case Reports

journal homepage: www.ajocasereports.com/

Surgical management of acanthamoeba chorioretinitis

Kapil Mishra [a], Gabriel Velez [a,b], C. Nathaniel Roybal [c], Vinit B. Mahajan [a,c,*]

[a] Molecular Surgery Laboratory, Byers Eye Institute, Department of Ophthalmology, Stanford University, Palo Alto, CA, 94304, USA
[b] University of Iowa Carver College of Medicine, Iowa City, IA, 52242, USA
[c] Retina Consultants of New Mexico, Albuquerque, NM, 87113, USA

ARTICLE INFO

Keywords:
Acanthamoeba
Chorioretinitis
Retina
Vitrectomy
Retinal detachment
Retinectomy

ABSTRACT

Purpose: Acanthamoeba chorioretinitis is a rare manifestation of the parasitic infection, and reported cases often result in enucleation. Surgical removal of Acanthamoeba chorioretinitis has not been previously described. We report a surgical case of Acanthamoeba chorioretinitis spread from keratitis that ultimately resulted in a disease-free outcome.

Observations: A healthy 80-year-old male with a history of keratoconus requiring a penetrating keratoplasty in the fellow eye presented with a severe corneal ulcer clinically consistent with Acanthamoeba keratitis. He ultimately required a penetrating keratoplasty and improved clinically until he developed vitritis on post-operative month 1 and was diagnosed with endophthalmitis. B-scan ultrasound demonstrated vitreous opacities and a large retinal mass that reduced in size following serial intravitreal injections of antibiotics, oral antibiotics, and a limited pars plana vitrectomy. He underwent a repeat pars plana vitrectomy 6 weeks later and a retinal mass in the mid-periphery with an associated tractional retinal detachment was noted. A localized retinectomy was performed around the lesion which was excised entirely, and silicone oil was instilled. Pathology of the lesion showed acute and chronic granulomatous necrotizing inflammation with the presence of several definitive amoebic organisms and numerous cells suspicious for amoebae. The patient was maintained on oral antibiotics by the Infectious Disease Service and was disease-free 1-year post-infection.

Conclusions and importance: Acanthamoeba chorioretinitis is a rare, devastating disease and often leads to enucleation. We present a surgical case showing control of the infection utilizing a surgical retinectomy. Aggressive local therapy and a multidisciplinary approach with the Infectious Disease Service may lead to a successful outcome.

1. Introduction

Enucleation is the most commonly reported treatment for Acanthamoeba chorioretinitis, a rare manifestation of the devastating parasitic ocular infection.[1–3] Typically infecting the cornea, Acanthamoeba ocular infection requires medical treatment with topical and often oral antiparasitic medications as the mainstay of treatment, although a corneal transplant may be required in severe cases. Despite aggressive intervention, ocular infections with Acanthamoeba frequently result in a poor prognosis as there often is a delay in diagnosis. If the infection extends into the posterior segment, the prognosis is worse, and the eye is often enucleated due to significant vitritis and retinal necrosis.[1]

Various regimens for Acanthamoeba keratitis have been extensively studied,[4] however to our knowledge successful treatment of chorioretinal disease has been rarely described. In this report, we describe a patient with Acanthamoeba endophthalmitis requiring a retinectomy with ultimately a disease-free outcome.

2. Case

A healthy 80-year-old male with keratoconus requiring a penetrating keratoplasty in the right eye at 36 years of age and repeat keratoplasty at age 74 presented with a severe corneal ulcer in the left eye which had not undergone a previous keratoplasty (Fig. 1A). His clinical presentation was consistent with acanthamoeba keratitis without an identifiable risk factor, and he ultimately required a repeat penetrating keratoplasty. The corneal pathology specimen confirmed amoebic cysts were present. After surgery, the patient improved until post-operative month 1 when he developed severe pain and dense vitritis. He was referred to the retina service for possible endophthalmitis. His visual acuity was hand motion

* Corresponding author. Byers Eye Institute, Department of Ophthalmology, Stanford University, Palo Alto, CA, 94304, USA.
E-mail address: vinit.mahajan@stanford.edu (V.B. Mahajan).

https://doi.org/10.1016/j.ajoc.2022.101388
Received 23 April 2021; Received in revised form 24 January 2022; Accepted 28 January 2022
Available online 31 January 2022

and the intraocular pressure was 5 mm Hg in the affected eye. Slit lamp examination demonstrated a clear corneal graft, mild anterior chamber inflammation, pseudophakia, and dense vitreous cell with no fundus view. B-scan echography showed dense, diffuse vitreous opacities and a localized retinochoroidal detachment measuring 4.4 mm in height with a dense subretinal opacity at the inferonasal equator (Fig. 1B). The patient underwent a 23-gauge pars plana vitrectomy in order to obtain a vitreous biopsy, debulk the vitreous debris, and deliver intravitreal vancomycin 1 mg/0.1 ml, ceftazidime 2.25 mg/0.1 ml, foscarnet 2.4 mcg/0.1 ml, and voriconazole 100 mcg/0.1 ml. A limited intraoperative view precluded a thorough examination of the retina. The biopsy showed rare cells suspicious for amoeba but no definitive organisms were identified, and most cells were consistent for mature lymphocytes (Fig. 1C). His postoperative course showed slow improvement in the vitritis and a reduction of the subretinal lesion to 1.9 mm on B-scan (Fig. 1D) after 3 intravitreal voriconazole injections over the course of 1 month, along with oral voriconazole 200 mg twice a day, acyclovir 400 mg 2 times a day, and doxycycline 100 mg 2 times a day. He was maintained on chlorhexidine drops 6 times a day, prednisolone acetate 1% 6 times a day, atropine 1% 2 times a day, and preservative-free artificial tears as needed in the affected eye.

Due to a significant post-intraocular lens opacity and persistent retinal mass with chronic low-grade inflammation, the patient underwent a repeat pars plana vitrectomy 6 weeks after the first vitrectomy. Intraoperatively, a tractional retinal detachment with extensive preretinal membranes was noted tracking to a retinal mass inferonasally (Fig. 2A). Membranes were peeled, and a limited retinectomy was performed surrounding the mass to relieve traction. The retinal mass was excised with the vitreous cutter and sent to pathology. An air-fluid exchange was performed to completely flatten the retina and endolaser was applied along the retinectomy border followed by silicone oil tamponade and intravitreal voriconazole 100 mcg. Postoperatively he was maintained on chlorhexidine drops 6 times a day, prednisolone acetate 1% 6 times a day, ofloxacin 4 times a day, atropine 1% once daily, oral voriconazole 200 mg 2 times a day, acyclovir 400 mg 2 times a day, and doxycycline 100 mg daily.

Pathological examination of the retinal mass biopsy demonstrated acute and chronic granulomatous necrotizing inflammation with the presence of several definitive amoebic organisms and numerous cells suspicious for amoebae (Fig. 2B and C). The Infectious Disease service recommended aggressive treatment with oral voriconazole 200 mg 2 times a day and trimethoprim/sulfamethoxazole (TMP/SMX) 2 double-strength tablets 3 times a day, however TMP/SMX was discontinued due to gastrointestinal symptoms and he was switched to oral flucytosine 2500 mg 3 times a day. The Infectious Disease service monitored complete blood count, creatinine, and liver function tests regularly. Oral flucytosine and voriconazole were discontinued by post-operative month-5 but restarted 1 month later due to an episode of scleritis concerning for recurrence. His symptoms resolved and there were no other episodes of inflammation for 1 year post-operatively while on voriconazole and flucotysine. The retina remained attached under silicone oil (Fig. 2D), and optical coherence tomography (OCT) revealed moderate cystoid macular edema that improved significantly by post-operative month 6 (Fig. 2E). Visual acuity remained stable at 20/200 with a clear corneal graft and no signs of recurrence at 1 year follow-up (Fig. 2G).

3. Discussion

We previously reported a patient with *Acanthamoeba* keratitis that inexorably progressed to sclerokeratitis and panophthalmitis requiring therapeutic enucleation.[1] In our current case, we were able to salvage the eye and isolate the area of chorioretinitis, which was confirmed by histopathology to contain definitive amoebic organisms. To our knowledge, this is the first reported case of Acanthamoeba chorioretinitis treated by surgical removal of the infectious mass and ultimately resulted in infection control, avoiding enucleation, and preservation of sight. A limited retinectomy has been well-described in Vitreoretinal surgery for severe ocular trauma,[5] however to our knowledge this is the first reported case of this surgical manuever being utilized for Acanthamoeba.

It is hypothesized that intraocular infection by Acanthamoeba is exceedingly rare due to a strong neutrophil response in the anterior chamber precluding intraocular spread.[6] In the rare cases of chorioretinitis, retinal whitening, hemorrhages, and even a subretinal abscess have been described.[1,2,7] Because of Acanthamoeba resistance to existing medications particularly in the cyst form,[8] treatment includes a combination of topical, intraocular, and systemic medication. Our case

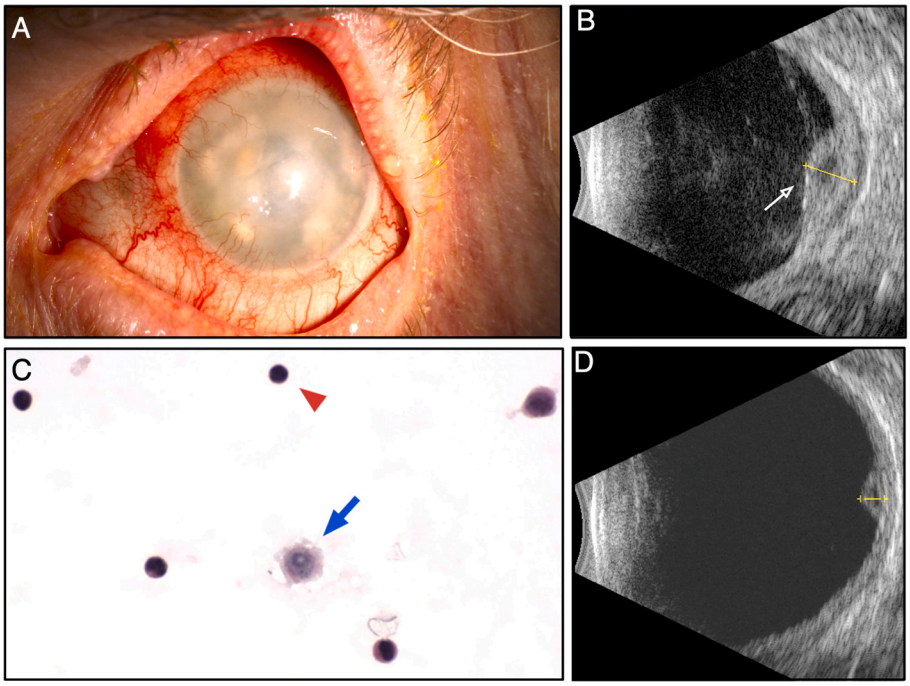

Fig. 1. Clinical imaging of acanthamoeba infection. **A**. Slit lamp photograph of the left eye demonstrates moderate conjunctival injection with ciliary flush and diffuse corneal haze with prominent corneal neovascularization. **B**. Preoperative B-scan ultrasonography showed moderate vitritis, choroidal thickening, and a chorioretinal mass (arrow) measuring 4.4 mm in height. **C**. Vitreous cytology suggested the presence of acanthamoeba trophozoites (blue arrow) and acute inflammation with lymphocytes (red arrow). Hematoxylin and eosin, original magnification 300x. **D**. After a core vitrectomy and intravitreal injection of broad-spectrum antibiotics, ultrasonography showed reduced vitritis, improved choroid thickening, and reduced retinal mass size. . (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

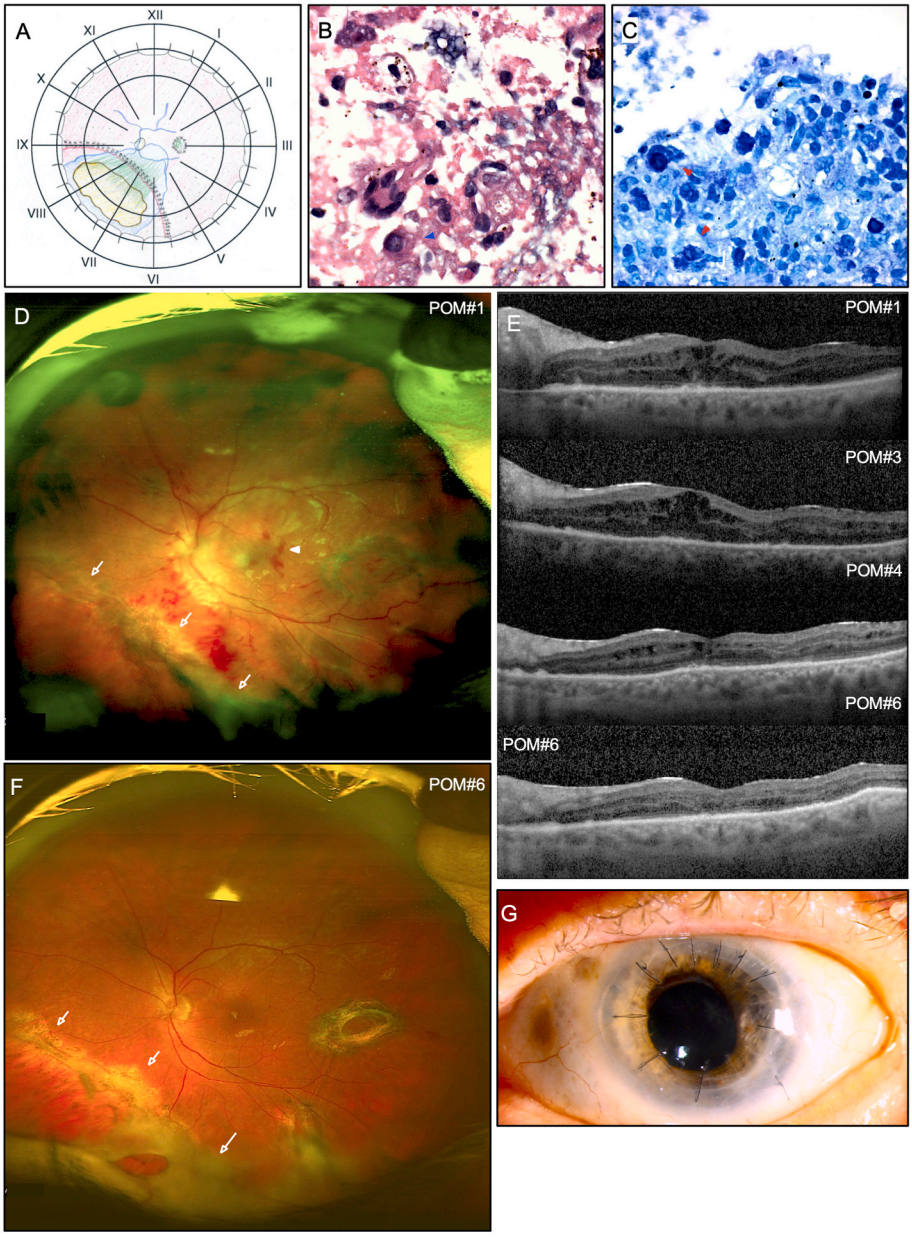

Fig. 2. Postoperative course after retinal biopsy where histopathological examination of retinal biopsy confirmed acanthamoeba cysts and trophozoites. **A**. Surgical drawing illustrates the inferonasal retinal mass in the mid-periphery with a tractional retinal detachment (blue) secondary to proliferative pre-retinal membranes (green). Pre-retinal membranes were also noted in the temporal macula. A large retinectomy to remove traction from the retinal mass was performed from 5:30 o'clock to 9:00 o'clock (red line) and the retinectomy edge was sealed with endolaser. **B**. Vitreous biopsy cytospin preparations showed lymphocytes, neutrophils and histiocytes. Rare cells had a morphology suspicious for amoeba (blue arrow). Hematoxylin and eosin, original magnification 300x. **C**. A retinal biopsy showed necrotic tissue with fragments of neurosensory retina and inflammatory cells including epithelioid histiocytes, lymphocytes, and neutrophils. Rare cells showed a morphology consistent with the trophozoite form of Acanthamoeba (red arrowheads). Wright-Giemsa stain, original magnification 300x. **D**. An ultra-widefield color photograph of the left eye on post-operative month 1 demonstrates attached retina under silicone oil. The retinectomy edge was flat (white arrows), and there was mild hemorrhage in the macula (white arrowhead). **E**. Spectral-Domain Optical Coherence Tomography of the left eye shows improvement of cystoid macular edema over the course of six months. **F**. An ultra-widefield color photograph of the left eye on post-operative month 6 demonstrates attached retina under silicone oil. The retinectomy edge remained flat (white arrows), and hemorrhages had resolved. No recurrence of retinitis was noted. **G**. Slit lamp photograph of the left eye shows a clear corneal graft with interrupted nylon sutures, and a quiet anterior chamber. . (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

suggests that vitrectomy surgery may play an important role by reducing infectious load by direct excision of infected tissue. In addition, co-management with the Infectious Disease Service helped guide appropriate systemic therapy and monitor side-effects during a long post-operative treatment course.

In conclusion, Acanthamoeba chorioretinitis is a severe manifestation of the parasitic infection and may benefit from vitreoretinal surgery. Close monitoring and aggressive treatment regimens are likely paramount to a successful clinical outcome.

Patient consent

Written consent to publish this case has not been obtained. This report does not contain any personal identifying information.

Funding

VBM is supported by NIH grants [R01 EY031952, R01 EY030151, R01NS98950, R01EY031360, and P30EY026877] and Research to Prevent Blindness (RPB), New York, NY, and the Stanford Center for Optic Disc Drusen.

Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Financial disclosure

None.

Declaration of competing interest

The following authors have no financial disclosures: KM, GV, CNR, VBM.

Acknowledgements

We thank Kenneth Goins for surgical care of the cornea and Nasreen Sayed for providing histopathology slides.

References

1. Mammo Z, Almeida DRP, Cunningham MA, Chin EK, Mahajan VB. Acanthamoeba endophthalmitis after recurrent keratitis and nodular scleritis. *Retin Cases Brief Rep*. 2017;11(2):180–182.
2. Moshari A, McLean IW, Dodds MT, et al. Chorioretinitis after keratitis caused by Acanthamoeba: case report and review of the literature. *Ophthalmology*. 2001;108 (12):2232–2236.
3. Davis MJ, Packo KH, Epstein RJ, et al. Acanthamoeba endophthalmitis following penetrating keratoplasty for Acanthamoeba keratitis. *Arch Ophthalmol*. 2010;128(4): 505–506.
4. Maycock NJR, Jayaswal R. Update on acanthamoeba keratitis: diagnosis, treatment, and outcomes. *Cornea*. 2016;35(5):713–720.
5. Ozdek S, Hasanreisoglu M, Yuksel E. Chorioretinectomy for perforating eye injuries. *Eye*. 2013;27(6):722–727.
6. Illingworth CD, Cook SD, Karabatsas CH, et al. Acanthamoeba keratitis: risk factors and outcome. *Br J Ophthalmol*. 1995;79:1078–1082.
7. Anderson NG, Hamler SE, Duncan LD. Primary subretinal abscess caused by acanthamoeba: clinical and pathologic case report and review of the literature. *Retin Cases Brief Rep*. 2012;6(1):37–39.
8. Coulon C, Collignon A, McDonnell G, Thomas V. Resistance of Acanthamoeba cysts to disinfection treatments used in health care settings. *J Clin Microbiol*. 2010;48(8): 2689–2697.